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**CASE REPORT**


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**Multilocular Cystic Renal Cell Carcinoma, Miss-Diagnosed Clinically as Renal Hydatid**
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**ABSTRACT:**

Multilocular cystic renal cell carcinoma (MCRCC) is a rare clear cell type of renal cell carcinoma. Although cytologically and cytogenetically MCRCC is similar to clear cell carcinoma, histology and behavior differ markedly. Prognosis in MCRCC is excellent as the tumour is of consistently low grade and surgical resection is curative. We report here a case of MCRCC in 44 year old female presented with right loin pain, was miss-diagnosed as renal hydatid cyst for 2years. Local excision of the tumor mass only was done with no recurrence after 30 months of follow up after. The differentiation between renal hydatid cyst and MCRCC may be difficult clinically and radiologically. The tumor grows very slowly. The outcome of the lesion is not affected adversely by large tumor size. There is an increasing belief that MCRCC should be renamed and reclassified to indicate much more favorable prognosis than the conventional clear renal cell carcinoma.

Key Words: Multilocular cystic renal cell carcinoma, clear cell renal cell carcinoma, renal hydatid cyst.

**INTRODUCTION:**

Multilocular cystic renal cell carcinoma (MCRCC) is a rare, usually unilateral tumor, with no side predominance<sup>(1)</sup> included in the 2004 WHO classification of renal neoplasms<sup>(2,3)</sup>. There is a male predominance of 3:1 with age ranging from 20 to 77 years<sup>(2, 3, 4)</sup>. The exact incidence of the lesion is unknown, reports vary between 1% and 14%<sup>(1,5,6)</sup>. The reason for this variability in reported incidence relates to the fact that diagnostic histological criteria have been inconsistent<sup>(7)</sup>.

Grossly, these tumors have a multilocular cystic appearance sometimes with evidence of a variegated, yellow solid component of limited size without expansive nodules or necrosis<sup>(1, 8)</sup>.

Histological criteria describe a well demarcated fibrous capsule housing cysts lined by flattened, cuboidal, clear cells and septa that contain aggregates of epithelial cells with clear cytoplasm. There must be no areas of necrosis present and any solid areas of clear cell neoplasm must occupy less than 25% of the total tumour volume<sup>(2,5)</sup>. Almost all cases have been cited as Fuhrman grades 1 and 2, with only rare incidences of T3 tumours<sup>(1,5)</sup>. The differential diagnosis of MCRCC includes multicystic kidney disease, segmental cystic disease, cystic nephroma, renal cell carcinoma with cystic change, intrinsic unilocular growth (papillary cystic adenocarcinoma), tumor arising from the epithelial lining of a pre-existing simple renal cyst and hydatid cyst.<sup>(9)</sup> A notable difference between MCRCC and conventional RCC is the absence of nodal or metastatic spread at diagnosis.<sup>(6,10)</sup> It seems that in general, cystic renal cell carcinoma has better outcome than non-cystic conventional RCC.<sup>(6,11)</sup>

Cytogenetic analysis has confirmed that 3p deletion is a frequent karyotypical abnormality in MCRCC<sup>[12, 13]</sup>. The 3p chromosome is known to harbour the von Hippel Lindau (VHL) gene (3p25). Interestingly, this gene has been implicated, along with other tumour suppressor genes located on the short arm of chromosome 3, in the progression of clear cell carcinoma<sup>[13]</sup>. This supports the likelihood that MCRCC is a subtype of clear cell renal cell carcinoma (RCC). The prognosis of MCRCC is excellent, after surgery with a disease specific survival rate is up to 100%<sup>(1, 14, 15, 16)</sup>. In the published literature to date, no MCRCC has ever recurred after surgical resection<sup>(5)</sup>.

We report here a case of MCRCC and discuss its clinical significance.

**CASE REPORT:**

A 44 year old female presented with right flank pain since October 2007. The pain was dull, coming on and off, of variable severity. She consulted a doctor at that time who diagnosed

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her as a case of renal hydatid cyst depending on ultrasonography and CT findings of presence of right renal hypo-echoic cystic mass of 106x100 mm of regular wall with multiple thin septation. He explored her transperitoneally in November / 2007 ,but he did do anything just aspiration of clear fluid from this mass .The patient was put on albendazole 400mg twice daily for three months with no improvement. In September / 2009 she was seen by one of the authors complaining of increasing pain with no significant findings in physical examination. Full work up was done for her including complete blood picture, blood biochemistry, renal function tests, and chest x-ray which were all normal .Unfortunately, serological tests for hydatid disease were not available in our locality. Abdominal ultrasonography revealed mixed echogenic mass of 115x108mm ,with multilocular cystic component. CT scan showed thick, regular wall, cystic mass with thin septae involving right kidney. There was no solid component , (Figuer-1a).Intravenous pyelography (IVU) revealed pushing of the kidney medially and down-ward with stretching and elongation of lower calyces .Exploration through retroperitoneal approach revealed: cystic mass of 120x109mm size, attached to anterior surface of right kidney with the bulk of the mass extending outside the kidney(Figure1b). The mass was well circumscribed and encapsulated by thick capsule, whitish capsule (Figure-2a). With these findings and a previous diagnosis of hydatid cyst; a decision was made to do a closed cystopericystectomy without opening of the mass as a cleavage line between the mass and renal parenchyma was easily dissected (Figure-2 b). There was no extension into perinephric fat or regional lymphadenopathy. The cut section of the mass revealed multiloculated, cystic lesion .The cysts ranged from 2 mm to 2.5 cm. The content of the cysts was clear serous fluid. There was no blood or clots or necrotic area . Histopathological examination revealed a multicystic tumor, lined by one layer or few layers of cuboidal malignant epithelial cells with clear cytoplasm and mild nuclear atypia.Small nests of tumor cells invade the adjacent renal stroma. The cysts were separated by fibrocollagenous connective tissue (Figure-3 a and b).The surgical resection margin was free of malignancy. After 30 months follow up with ultrasonographic examination each 3 months and

CT scan yearly, the patient is well with no recurrence.

### DISCUSSION:

MCRCC appears to be a distinct subtype of RCC, with characteristic gross and microscopic features <sup>(8)</sup> and appears to have a favorable biology.<sup>(9)</sup>In 1957,Robinson described the first case of so-called MCRCC.<sup>(17)</sup>No tumor with these features has ever recurred or metastasized<sup>(1,2)</sup> ,except one reported case <sup>(7)</sup>. Therefore, the use of these somewhat more restrictive criteria to diagnosed MCRCC should be adopted in daily practice.

In this case; First : It is clearly that the histopathology was diagnostic of multilocular cystic renal cell carcinoma depending on the characteristic gross and microscopic criteria in the 2004 WHO classification.<sup>(2)</sup> During the last 19years, the author had done 98 radical nephrectomy for patients with RCC beside this case of MCRCC. So, it accounted for 1.0% of all cases. This is similar to the world reported incidence of this subtype.<sup>(6)</sup>

Second: Nuclear grade was determined according to the Fuhrman system<sup>(18)</sup>,and tumor stage was assessed according to AJCC cancer staging manual 2010.<sup>(19)</sup>The tumor stage in our case was T2bN0M0 and of Fuhrman grade 2. The size of the tumor in this case was relatively large at diagnosis due to delay of proper diagnosis for about 2years. Moreover, the tumor grows very slowly as it was increasing in size by only 9x8mm in these 2years depending on radiological assessment.

Third: On ultrasonography and computed tomography images, MCRCC appeared as a well-defined, multilocular cystic mass containing fluid, with no expansile solid nodules in the thin septae and sometimes with small, slightly enhanced solid areas.<sup>(20)</sup>When radiological examinations demonstrate a cystic renal mass of this kind in adult, renal hydatid cyst should be included in the differential diagnosis especially in endemic area with hydatidosis like Iraq , as we misdiagnosed our case. Also the absence of specific symptoms and signs in both renal neoplasms and renal hydatid cyst make the differentiation between them very difficult.

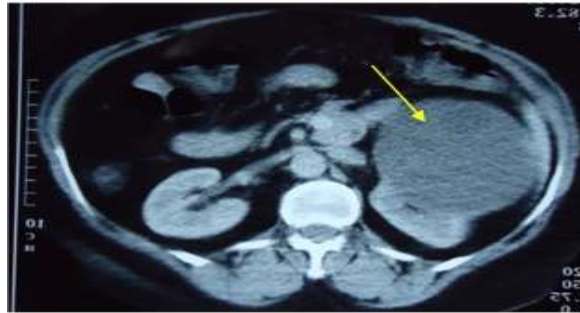
Fourth: In spite of the inaccurate pre and intra-operative diagnosis, we performed local excision of the tumor mass, with no recurrence after 30 months of follow up. This indicates the low malignant potential of this tumor and need for

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minimal nephron sparing surgery whenever technically feasible. This could be achieved if the accurate diagnosis of MCRCC was done preoperatively. But, this may be difficult.<sup>(21)</sup>Pre-operative biopsy and frozen sections may not

always diagnose the lesion with certainty.<sup>(21)</sup>Magnetic resonance imaging with strict CT criteria may make the preoperative recognition of MCRCC possible. This needs further studies on large number of cases.



**Figure (1a) CT shows a cystic mass at the upper outer Part of the right kidney**



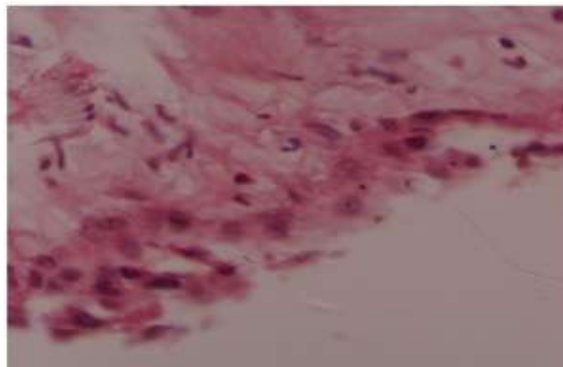
**Figure (1b) Intraoperative photo showing a cystic renal mass**



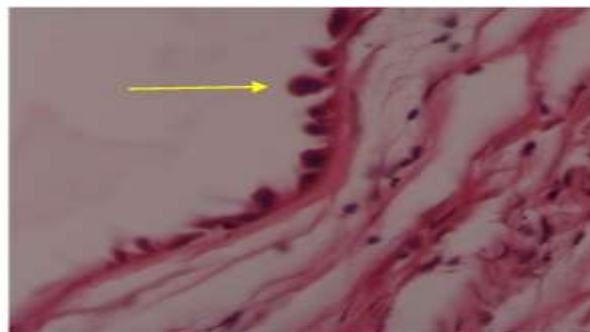
**Figure (2a) A multilocular cystic mass with thick capsule**



**Figure (2b) A multilocular renal cystic mass on sectioning**



**Figure (3a) A microscopic photograph showing Malignant epithelial cells within the stroma  
Original magnification X400**



**Figure (3b) A microscopic photograph showing malignant cuboidal cells lining the cyst  
Original magnification X400**

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